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Case Report

DIAGNOSTIC APPROACHES TO MOYAMOYA DISEASE IN A PATIENT WITH HEAD TRAUMA

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ABSTRACT

Introduction: Moyamoya Disease (MMD) is a chronic cerebrovascular disorder characterized by progressive stenosis and occlusion of the distal internal carotid artery and the proximal segments of the middle and anterior cerebral arteries. This leads to the development of abnormal vascular networks at the base of the skull, resulting in a "puff of smoke" appearance, which is a hallmark of the disease[1]. This condition leads to reduced cerebral blood flow and a heightened risk of stroke. Early diagnosis through imaging techniques like Digital Subtraction Angiography (DSA) is essential for effective management.
Case Report: A 27-year-old male presented with decreased consciousness after a traffic accident. He was initially assessed with a Glasgow Coma Scale (GCS) score of E1M2V1 and left-sided hemiparesis. Despite undergoing decompressive craniectomy, the patient developed left-sided hemiplegia one week later, which did not correlate with the initial haemorrhagic pattern on CT. DSA revealed stenosis from the C7 segment to the A1-M1 proximal right ICA, and the case was classified as Suzuki stage IV.
Discussion: MMD diagnosis relies on advanced imaging, with DSA being the gold standard for assessing the extent of stenosis and collateral vessel formation. The Suzuki classification system helps determine disease progression and informs treatment decisions. In this case, DSA provided crucial information for diagnosis and management, highlighting the advanced stage of the disease.
Conclusion: This case underscores the importance of early diagnosis in Moyamoya disease, particularly using DSA for accurate staging. Timely intervention is critical to reduce stroke risk and improve outcomes, especially in advanced stages.

Keyword: *diagnostic; dsa; head injury; moyamoya*

INTRODUCTION

Moyamoya disease (MMD) is a chronic cerebrovascular disease, characterized by progressive stenosis and obstruction of the distal portion of the internal carotid artery and proximal portion of the middle and anterior cerebral arteries, resulting in the development of abnormal vascular tissue at the base of the skull, resulting in a smoke-like or moyamoya appearance[1]. Moyamoya patients can be divided into two groups, namely patients with primary moyamoya disease (MMD) and patients with secondary moyamoya caused by other known etiological factors, such as intracranial atherosclerosis (moyamoya syndrome [MMS]).

The incidence rate of MMD is very high in East Asia, especially Japan and South Korea, compared to other countries and is especially common in women. Moyamoya disease has two peak ages of onset: between 5 and 9 years, and between 45 and 49 years[2]. MMD represents a variety of cerebrovascular events including TIA, ischemic stroke, intracranial hemorrhage, headache or seizure[3]. MRI of the brain and arteriography are imaging modalities that allow a positive diagnosis of this disease.

CASE REPORT

The patient was a 27 year old male who was unconscious after a traffic accident. There was no vomiting during the incident, both ears and nose were bleeding. On physical examination, the Glasgow Coma Scale score was E1M2V1, a fracture on the right side of the face accompanied by hemiparesis on the left side. The patient underwent a decompressive craniectomy and 1 week later there was left-sided hemiplegia which did not match the location of the bleeding based on CT scan patient's head.

The results of the CT angiography supporting examination were impressive suspected thrombosis of the right distal internal carotid artery with stop filling start segment opthalmic right with the anterior cerebral artery and the right middle cerebral artery from proximal to distal appearing to be supplied by the left internal carotid artery via the anterior communicating artery.

To further substantiate the diagnosis, additional imaging modalities, including Digital Subtraction Angiography (DSA), were performed. The DSA findings revealed ICA stenosis in the RCCA segment from multiple projections. The Acomm artery was not visualized, and an anastomosis between the middle meningeal artery (MMA) and a branch of the ICA was observed, indicating stenosis of the ICA from the C7 segment to the proximal right A1-M1.

Other supporting examinations carried out are: Trans Cranial Doppler (TCD) with a conclusion low flow velocities with high resistance flow detected at right ICA it may suggest right ICA near/total occlusion. The diagnosis in this patient was made based on the Suzuki classification and through supporting examinations in the form of CT angiography, DSA and TCD. For this case, the patient is classified as Suzuki classification stage IV.

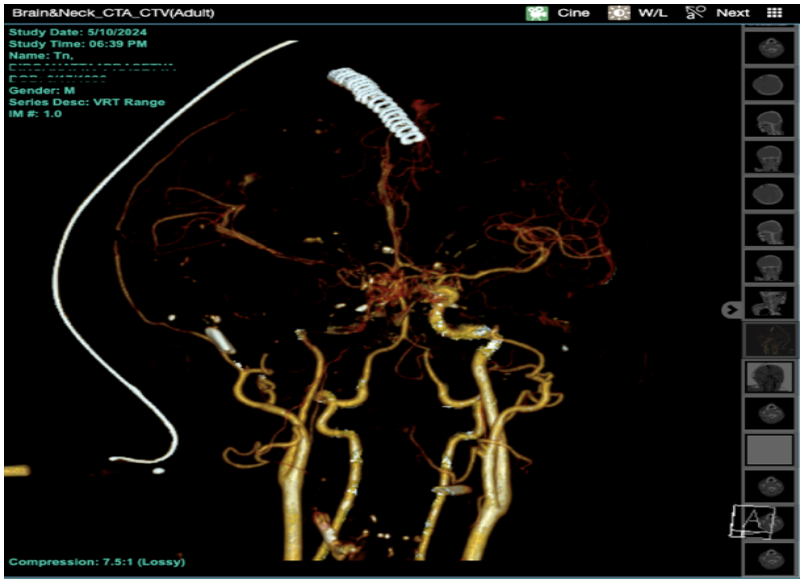

Figure 1. CT Angiography

Artery	Velocity (cm/sec)							
	LEFT				RIGHT			
	Systolic	Diastolic	MV	PI	Systolic	Diastolic	MV	PI
Internal Carotid Proximal	93	34	53,67	1,10	38	3	14,67	2,39
Internal Carotid Distal	82	33	49,33	0,99			0,00	0,90
Bulb	89	32	51,00	1,12	60	11	27,33	0.85
External Carotid Proximal	70	19	36,00	1,42	84	17	39,33	1,70
Common Carotid Proximal	105	31	55,67	1,33	93	9	37,00	2,27
Common Carotid Distal	88	29	48,67	1,21	67	17	33,67	1,49
Vertebral Mid	46	17	26,67	1,09	42	12	22,00	1,36
Vertebral Origin	45	11	22,33	1,52	40	12	21,33	1,31
ICA/CCA Ratio								

Figure 2. Transcranial Doppler Result

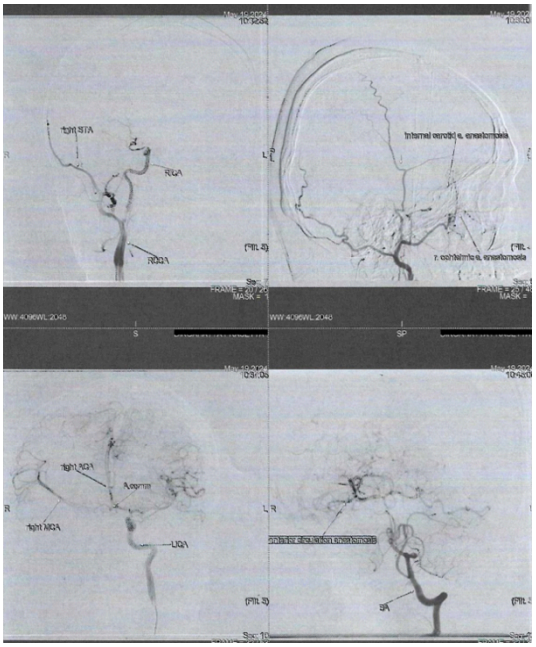

Figure 3. DSA of Internal Carotid Artery

DISCUSSION

First-line imaging studies include CT angiography and MR angiography to identify anatomical characteristics, such as steno-occlusive changes and collateral vessel formation, as well as *digital subtraction angiography* (DSA)**,** which continues to be the gold standard examination for confirming diagnosis and determining classification because it has high spatial and temporal resolution[4].

However, CTA has limitations in depicting small caliber micro vessels (MMV) (<3 mm) and has lower resolution than DSA. Additionally, CTAs can also be disrupted by motion artifacts[5]. The gold standard for angiographic diagnosis of MMD is the Suzuki grading system.

The Suzuki classification system describes the process from initial stenosis of the terminal part of the internal carotid artery (ICA) to the appearance of deep but fragile collateral tissue, as well as subsidence of the moyamoya vessels accompanied by the simultaneous development of blood supply from the branches of the external carotid artery. This fragile collateral tissue mainly develops from the thalamoperforating arteries and lenticulostriate perforating arteries, where the cerebral angiography findings of MMD patients are divided into 6 stages[6] (table 1).

This stage is very important for determining the prognosis and treatment options for patients with Moyamoya, especially regarding the development of collaterals and the risk of complications such as ischemic stroke or hemorrhage.

Steno-occlusive changes in the ICA terminus region and subsequent development of 'moyamoya' vessels become the main features[7].

DSA is able to clearly identify the degree of stenosis or occlusion of blood vessels, as well as reveal the presence of abnormal neovascularization at the base of the brain. In addition, DSA also provides an accurate evaluation of the pattern and hemodynamics of compensatory blood supply[8].

Conventional cerebral angiography or DSA is still considered necessary for the definitive diagnosis of Moyamoya, although this procedure carries a risk of complications[9].

Access to healthcare facilities that provide DSA may be limited, and the invasive nature of the procedure may prevent some patients from undergoing testing. The importance of accurate diagnosis of Moyamoya disease in affected patients has become increasingly clear through advances in catheter angiography techniques and the emergence of CT and MRI technologies[10].

(EDAS) and *encephalo-myo-synangiosis*

Stage	Angiography Findings
I	Narrowing of the carotid bifurcation On angiography examination, only the terminal part of the internal carotid artery (ICA) is stenotic or narrowed. This stage represents the beginning of the disease process with limited disruption of the main arteries without significant collateral formation.
II	Initiation and emergence of basal Moyamoya On angiographic examination, stenosis was seen in all terminal branches of the ICA (anterior cerebral artery [ACA] and/or middle cerebral artery [MCA]), and the deep Moyamoya vessels were beginning to appear. Collaterals begin to form in the basal areas of the brain, and these blood vessels become fragile and susceptible to bleeding.
III	Increased basal Moyamoya On angiographic examination, the deep Moyamoya blood vessels become more visible or intensive. Magnetic Resonance Angiography (MRA) at this level shows the picture "puff of smoke" which is typical. There is a deflection or change in direction of the ACA and MCA, which indicates a disturbance in blood flow.
IV	Shrinkage of basal moyamoya On angiographic examination, the deep Moyamoya blood vessels begin to experience regression or shrinkage, while transdural collateral blood vessels begin to appear. Deflection in the posterior cerebral artery (PCA) begins to be noted at this stage, indicating compensatory blood flow through the new pathway.
V	Moyamoya reduction On angiography, there was further regression of the deep Moyamoya vessels, and progression of transdural collateral vessels that replaced most of the blood flow. These blood vessels continue to grow to maintain blood supply to the brain.
VI	Disappearance of moyamoya On angiographic examination, the deep Moyamoya vessels had completely disappeared, and there was complete occlusion of the ICA. The blood supply to the ACA and MCA areas comes primarily from the branches of the external carotid artery, which is the main route for cerebral blood flow at this stage.

Table 2. Suzuki Staging in Moyamoya Disease

Operative measures such as revascularization play an important role in the management of patients with Moyamoya disease to reduce the number of ischemic and hemorrhagic strokes. Various revascularization strategies to reduce ischemic impact have been previously reported; these strategies include direct bypass, such as superficial temporal artery (STA)–MCA anastomosis and STA–ACA anastomosis; indirect bypass, such as *encephalo-duro-arterio-synangiosis* (EMS), as well as a combination of direct and indirect anastomosis[11].

The overall prognosis of Moyamoya disease is variable. About two-thirds of patients with Moyamoya disease experience progression of symptoms within 5 years with poor outcomes. The occlusion process continues regardless of the severity of symptoms, current treatment, age, gender, type and location of disease[11].

CONCLUSION

In this patient with a case of head trauma, the diagnosis of Moyamoya disease was confirmed through diagnostic examination using DSA and was classified as Suzuki stage IV. DSA is considered the gold standard in the diagnosis of Moyamoya disease with a high success rate in identifying typical arterial stenosis and collateral vessel formation.

DSA should be performed as early as possible in clinically suspected cases. In addition, DSA is the best choice to observe the formation of collateral circulation which is very important in determining the treatment strategy and evaluating the status of neo-angiogenesis after surgery.

However, considering that access to health services in Indonesia is still limited, both in terms of geography, facilities and distribution of human resources, this is a challenge in implementing it widely

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